

White (J. C.)

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of James C. White.*

FOUR CASES
OF
SCLERODERMA.

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PROFESSOR OF DERMATOLOGY IN HARVARD UNIVERSITY.





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SCLERODERMA has attracted the attention of dermatologists so much in the past few years, and so good descriptions of the disease are to be found in their writings, that nothing need be said in introduction concerning the following cases. I have been led to report them because the affection is a very rare one and its nature needs further investigation, and because the additional data they furnish are in some respects novel.

Case 1. — E. M. G., the patient, was first seen by me in June, 1874. She was a girl 22 years old, and her health had always been good until two years previously, when it gradually failed without apparent cause, or more definite symptoms than a little indigestion. She continued at her employment in the chemical chamber of a photographer for six months, when for want of strength and ability to move her limbs freely she gave up work, and has done none since. It was at this latter period that her attention was first drawn to the peculiar change in her condition. She noticed that her hands and knees were of a darker color than the rest of her skin, that she could not use her fingers well, and that her knees were so stiff that she could not easily raise herself to a standing position when sitting. From this time, for a year or little more, the changes in her skin very gradually progressed to the state in which I first saw her, but there had been no marked advance in them for the three or four months immediately preceding my examination.

The face presented a dark appearance over its lateral regions, and as the patient was a blonde, the melasma was very noticeable. But what especially attracted attention was the placid, immovable look she wore during conversation or laughter, as if she had never learned to use her facial muscles. She could wrinkle her forehead slightly, and could close her eyelids without much difficulty. The movement of the mouth and jaws, however, were considerably impaired, and she couldn't whistle, although she "used to like to." The whole integument of the face felt hard and thickened, the induration being greatest over the forehead and sides. The ears were somewhat stiffened. The neck and bust were very dark colored,

and the whole front upper chest felt hide-bound and was marked by numerous small white streaks, the pigment in these linear districts in reality remaining in normal quantity, but by contrast giving the appearance of parallel lines of leucoderma. The breasts were unaffected. The arms were very dark colored throughout, and below the elbows the skin was very dense. The movements of the elbow-joint were considerably impeded. The hands were very dark, resembling those of a mulatto, and running along the backs of the fingers were seen several longitudinal white streaks like those upon the chest. It was seen here more plainly than in the latter region, that these lines were made up of white points corresponding to the mouths of the follicles and sweat-glands, around which the melasma had failed to develop itself. The skin of the hands was very firm, so that they felt like those of a statue, and only a very slight motion of the phalangeal joints was possible. The thighs were very dark colored, but with the exception of the region of the knees, around which the skin was considerably thickened and hardened, the integument of the lower extremities was not much changed in consistence. The toes were scarcely at all affected.

The functions of the skin appeared unaltered, although in the beginning she thought the perspiration was for a time lessened over the whole surface. The cutaneous temperature was possibly a little lowered, but not materially so, and the sensibility of the skin to touch and pain was unimpaired. There had been but little change in her weight, and the state of her general health was satisfactory in all respects, and had it not been for her hide-bound condition, especially of the upper extremities, she would have been able to work.

Four months after the first examination the patient was again seen. She had been in good health, and there had been apparently no advance in the progress of the disease.

In April, 1875, I saw her again. She had been suffering greatly from indigestion, so that she had lost nearly twenty pounds of flesh, but little alteration in the condition of the skin was noticeable. She had had "sores" upon the knuckles and tips of the fingers, suppurating and lasting several weeks, the result of bruises and knocks, she said. The marks of these were plainly visible. The color of the hands was even darker than before, and several leucodermic patches of considerable size were for the first time seen upon the backs in strong contrast with the general melasma of the parts. Her condition is well expressed by her remark at this time, that persons called at her house and asked to see "the petrified girl."

Case 2.—Mrs. C. B., æt. 38. This patient was seen in November, 1873. She was born in Ireland, and had always been well until the

previous January, when, after taking cold, her catamenia suddenly ceased and never reappeared. Soon after this she felt prickling and twitching sensations in her face and arms, with "drawing feelings," and noticed that the skin of these parts was changing color and becoming hard. At the same time she began to lose flesh and spirits. In the following May she was married for the first time. Her general ill health and the affection of the integument had gradually progressed to the condition presented at the date of examination.

At that time the patient was considerably emaciated and reduced in strength. The skin of the whole face was very firm, so that the movements of all the facial muscles were made with difficulty. Its color was generally dusky. She had difficulty in eating on account of the induration of the integument about the mouth, and her tongue seemed larger to her than formerly, although it was not changed appreciably to touch. The face presented over the upper parts a fine papular, follicular inflammation, which had preceded, and had not been modified by the development of the sclerous condition. There was also an increased sebaceous flux over the face, so that the combination of this glossiness with the general melasmic hue, and red tints of the follicular hyperæmia gave to the immobile and stony countenance a most peculiar aspect. The arms from the elbow downward were of a dusky color, the pigment deposit growing deeper to the finger ends. A similar progression in intensity downward marked the development of the hardening of the integument over the same parts, the skin of the hands, especially the fingers, being seemingly converted into sole leather, so that motion in them was very difficult. The skin of the upper portion of the trunk was deeply colored, and the respiratory movements of the chest were considerably impeded by the inelastic induration which had taken place in it. Upon the lower extremities only a slight degree of hardening was noticed, and there was but little difficulty in walking.

The hands felt cold to the patient at times, but there was nowhere any great change in the temperature of the skin or in its sensibility. The sclerosis was decidedly more marked upon the right arm and leg than upon the opposite half of the body.

The patient was seen several times subsequently during a period of six months, but little change in the condition of the skin was noticed. Her general health, however, deteriorated, and symptoms of pulmonary disease were making themselves manifest. Of her condition during the past year, or if she be still alive, I have no information, as she lived in another city.

Case 3.—C. E. This patient was a woman 54 years old, and a native of Ireland. Her health had been generally good until two months before I saw her (December, 1874). At that time she had

had a severe and sudden attack of pain in the region of the left axilla, which was followed in a few hours by swelling of the parts. How long these symptoms lasted it was impossible to learn positively, but probably a few days. Very soon afterward she noticed a change in the left hand, which continued to increase until she consulted me concerning it.

A careful examination of the axillary and neighboring regions failed to discover any tissue change there. The left hand, however, presented this very peculiar condition. The skin of the ulnar half was marked by irregularly shaped, faint melasmic stains upon the dorsal surface, and was very dense and firm. Upon the third and fourth fingers the change was still more apparent. They felt as if made of wood, and could only be flexed about one-half the usual extent. A very slight induration of the skin about the wrist upon its ulnar side was apparent, but further than this no change was detected. The other fingers and thumb were in their normal state.

Case 4.—T. G. is a Scotchman, 42 years old. He lived in Glasgow until the age of 23, when he enlisted in the marine artillery, and served in the Baltic two years during the Russian war. He returned to Scotland, but eighteen months subsequently came to this country, and has worked in a neighboring town as a stone mason, with the exception of one year's service in our cavalry during the war of the rebellion, at which time he had "chills and fever." This was his only sickness until the beginning of his present trouble. I first saw him in February, 1874.

Eighteen months previously he noticed that the skin over large portions of his body was covered with small white elevations, and looked, he says, like that of a plucked goose. These prominences soon disappeared, he thinks, but the skin afterward continued to feel tight and bound. Six months afterward his forehead began to assume its present appearance, and severe pains were felt extending from the elbows to the fingers, which lasted two months and ceased, leaving the skin of the hands and forearms tight. Six months before I saw him, the integument from the right shoulder to the top of the head upon the same side became inflamed and covered with blisters. This attack lasted three weeks, and was called by the family erysipelas. A physician who met him said he looked as if he had been poisoned by dogwood (*Rhus*). During the last six months before I saw him, he thought there had been but little change in the state of the skin. This was as particular a history of his previous condition as he could give.

On inspection the face seemed, at first, to be marked by an extraordinary development of the frontal protuberances, by a very broad nose, and to be very strongly pitted over its upper part by small-pox. Its general hue was slightly redder, and it was more glossy

looking than was natural. The eyes appeared to be only half open, and as if hidden by the overhanging prominences above. On closer examination the skin of the forehead was found to be intensely hard and greatly thickened, and to be marked by depressions varying in size from a pin's head to a pea. Over the frontal sinuses were broad and dense projections, and between them, running down to the nose, a rigid, elevated, semi-cylindrical band of cicatricial hardness, the surface of which was marked by the same peculiar depressions. The nose in its upper half broadened out and merged itself into the cheeks, the skin of the parts being of an almost bony hardness. From the alae there radiated to the lower cheeks lesser bands of the same cicatricial hardness, which were not elevated above the general surface. The skin surrounding the mouth was also very dense. He could not move his face to laugh, and he had not been able to whistle or play the flute for a long time. He had some difficulty even in getting his food into his mouth. The ears were very stiff and large, and were everywhere marked by small and large prominences of intense hardness. Surrounding them in all directions, even upon the scalp nearly to the vertex, there were seen still more exaggerated and abruptly defined elevations of cartilaginous feel and white color, which formed with the interspersed depressions of a deeper color a very striking contrast. The trunk was quite generally covered on its front and back surfaces with thickly clustered papular elevations, varying in size from a pin's head to a small pea, which were white, and looked like over-filled sebaceous glands. They were of intense hardness, however, to the touch, and cut like gristle. On the upper chest these elevations were arranged in parallel rows, as if occupying the seat of excoriations produced by the nails in scratching. The skin of the arms, beginning half way above the elbows, was of increased hardness, and more dusky in color than elsewhere. Below the elbows the induration increased progressively to the finger-tips, the hands being very hard as if encased in sole leather, but the surface of the upper extremities was perfectly smooth and free from the elevations and depressions so conspicuous elsewhere. Scarcely any movement of the fingers was possible, and work had been given up eight months previously from inability to hold the drill. The color of the hands was paler than natural, excepting between the fingers, where a dusky hue was noticeable. The thighs showed some general hardening of the skin, and at the upper part an occasional group of small papular and linear ridge-like elevations similar to those upon the trunk.

The sensibility of the skin was in no way blunted, and its temperature was nowhere affected except upon the hands. These felt cold to him, and were somewhat over-sensitive on pressure. They did not perspire, moreover, to any appreciable amount, although

the action of the sweat-glands elsewhere was unaltered. He felt generally hide-bound, and the skin seemed more sensitive to cold than formerly. His general health was very good, although he did "not feel quite so strong as he used to." His functions were all in good order, and had it not been for the immobility of his arms and hands he could have worked nearly as well as ever.

In the beginning of 1875, just one year after the above description was taken, he was again examined. His health had remained uniformly good. There was but little change in the condition of the skin; possibly the appearances were a little more pronounced, certainly there had been no retrogression. His only complaint was of an itching about the genitals, and a few minute excoriations from scratching were seen about these parts. Upon the lower abdomen and the upper femoral regions it was noticed that small, papular, firm and colorless elevations were beginning to show themselves arranged in parallel rows, together with prominent linear ridges of the same dense consistence. The latter were much more marked when the skin was forcibly stretched, and their position and arrangement reminded one of the marks of linear atrophy in women, except that here we had elevations instead of depressions.

In April the patient was again seen. He was recovering from an attack of pneumonia, which had left him considerably reduced in flesh and strength, but had been followed by no marked change in the condition of the skin. A portion of one of the nodular masses behind the ear was removed, and its structure examined by the microscope. The epidermal layers were normal in appearance; possibly a little thickened. The papillæ were somewhat less prominent than natural, and they, as well as the rest of the integument below, as far as the incision extended, were converted into a dense structure consisting of a firm network of fibrous tissue. The elastic fibres were very numerous. But few cell elements were seen, and no glandular structures were seen in the sections examined. Dr. Fitz, who also examined the specimen, found "no evidence of enlarged lymphatics," and likened the tissue unto that of elephantiasis arabum.

The first two of the above cases may be regarded as typical in their course and appearances, while the last deviates so widely, in some respects, from cases hitherto recorded, that I hesitate to class it among them, and do so, partly, because its position elsewhere among recognized affections of the skin would be still more inappropriate.

In all of them we fail to detect any exciting causes of the development of the disease, and the constant good health, prior to the

local affection, is especially noticeable. Three of the patients were women; one a man. The ages at which the diseases began were respectively 21, 37, 54 and 41. Two were of Irish descent, one was Scotch, and one an American. The most remarkable feature common to all, in addition to the sclerosis of the integument—the one essential element of the disease—was the pigment change expressed by melasma, and, to some slight degree, by leucoderma of the parts affected. The functions of the skin were but slightly modified, the sweat glands acting with a little irregularity in some cases, while there was a slight degree of seborrhœa in two and an acne in another of them, upon the parts affected. In all four, the upper parts of the body were most seriously affected, the sclerosis being most highly developed upon the fingers. A want of lateral symmetry was shown in cases II. and III., the affection being most pronounced upon the right half in the former, whilst in the latter it was not only confined to the left hand but to the ulnar side of this. In connection with the opinion founded on the pathological anatomy of the disease—that it is primarily an affection of the lymph system, and that the new formation of fibrinous tissue of which it essentially consists is analogous to the process in elephantiasis arabum—it is interesting to note the occurrence of an inflammatory process of the skin in the last two cases. In number III it is fair to conclude that this attack, which immediately preceded the sclerosis, was, in some direct way, connected with its development; but in the last case the inflammation of the integument, although it may have been of an erysipelatous character, did not come on until a year after the beginning of the general affection, and was confined to a small part of the affected regions, so that it can hardly be regarded as possessing any more intimate connection with the disease than that of an accidental occurrence. In fact, the whole clinical history of scleroderma indicates that, although closely allied to elephantiasis Arabum in its anatomy, it is wholly unlike it in etiology.

I can nowhere find mention of a case like this last in the descriptions of scleroderma or other affections of the skin. Dr. Duhring and Dr. Wigglesworth examined it with me during the past winter, but neither of them, I believe, had ever seen its like. It is unlike scleroderma in the marked depressions and elevations which form

its surface appearances, although Kaposi describes prominent, cord-like indurations in two cases, which suggest to him the identity of the disease with Addison's keloid. The pointed nodules about the ears, the tubercular prominences about the neck, and the broad and elevated bands or ridges on the forehead, taken by themselves remind one, in fact, most strongly of some forms of keloid and hypertrophied scars. The pigment alterations, moreover, were less developed than ordinarily in scleroderma. In its seat, however, its course, and the nature of the tissue changes which determine its true character, the affection is identical with scleroderma, and the condition of the skin of the forearms and hands differed in no way from the ordinary manifestations of the disease. The case might properly be called scleroderma, with an exceptional tendency to exuberant outgrowth.

BOSTON, *May*, 1875.

